The site of the lesions in “vestibular neuritis”

Murofushi et al. studied the site of lesions in “vestibular neuritis” using galvanic vestibular evoked myogenic potentials. Among the 11 patients diagnosed with vestibular neuritis, eight showed a neuritis pattern whereas three showed a labyrinthitis pattern.

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Commentary by Michael Halmagyi and James Colebatch

It is over a decade since Neurology published the first report of a short latency vestibulo-collic reflex we called the vestibular evoked myogenic potential (VEMP), recordable from anterior neck muscles, specifically the sternomastoid, in response to loud clicks. Since then, the VEMP has been studied in vestibular laboratories worldwide and has been shown to have application in the assessment of a range of vestibular disorders, including superior semicircular canal dehiscence, Ménière disease, multiple sclerosis, brainstem infarction, vestibular neuritis, and vestibular schwannomas. The definition of this vestibulo-collic reflex pathway has allowed the development of other novel methods of vestibular activation such as head tapping, mastoid bone vibration, and the technique used in this article, short duration galvanic (DC) currents. A galvanic current applied to the mastoid directly activates vestibular nerve endings. Murofushi and coworkers have produced much of the new clinical work in the last 5 years on VEMP. They previously reported that galvanic VEMP can separate vestibular nerve (retro-labyrinthine) lesions from vestibular end-organ (labyrinthine) lesions. They now turn their attention to defining the level of the pathology in vestibular neuritis. Sudden, isolated, total or subtotal, unilateral loss of vestibular function can occur during viral infections such as mumps and herpes zoster and perhaps herpes simplex. As a result, this form of unilateral loss of vestibular function has, like sudden unilateral loss of facial nerve function, been attributed to the direct or indirect effects of a viral infection of the vestibular nerve, “vestibular neuritis.” But, perhaps unsurprisingly, there is little pathologic evidence of neuritis in the vestibular nerve. Some patients with vestibular neuritis develop benign positional vertigo, a syndrome that actually indicates end-organ involvement. Nonetheless, uncertainty remains as to the exact site or sites of lesion.

In this issue of Neurology, Murofushi et al. present click and galvanic VEMP in patients with severe vestibular neuritis, studied within a month of onset, who had all lost the click VEMP on the affected side. They found that whereas most had also lost the galvanic VEMP, implying involvement of the vestibular nerve, three had preserved galvanic VEMP, implying that the vestibular nerve remained intact. This indicates that at least in some patients, the lesion is primarily in the labyrinth. Although their results show that most patients with vestibular neuritis have evidence of impairment of the vestibular nerve, even this could be an “upstream” effect. The additional involvement of the end organ—the labyrinth itself—cannot be excluded. It is not clear therefore whether these two patterns are indicative of two different types of pathology or part of a continuum. In light of their new results, however, the rather ungainly term “vestibular neuro-labyrinthitis” seems more accurate than “vestibular neuritis.”

References


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**A2A receptor inhibition in Parkinson’s disease**

Pharmacologic blockade of striatal adenosinergic A2A receptors has been proposed as adjunctive treatment of Parkinson’s disease. In a randomized, blinded study, Bara-Jimenez et al. found that the A2A antagonist KW 6002 potentiates and prolongs the antiparkinsonian action of levodopa, but with less severe dyskinesia.

**Neuropathology in restless legs syndrome: Further evidence for abnormal iron metabolism**

In a neuropathological study of individuals with primary RLS, Connor et al. found that transferrin receptor expression on neuromelanin cells is less than expected given the profile of iron deficiency in the substantia nigra. No other histopathology was observed in the restless legs syndrome brains.

**Randomized trial of istradefylline in advanced PD**

Hauser et al. report results of a 12-week, blinded, randomized, placebo-controlled, exploratory study of istradefylline in PD subjects with both motor fluctuations and peak-dose dyskinesias. Istradefylline was generally well tolerated and reduced OFF time as assessed by home diaries.

**Cycad neurotoxins are biomagnified in Guam flying foxes**

Banack and Cox report that flying foxes from Guam have extraordinarily high levels of the cycad neurotoxin BMAA. This finding is consistent with their hypothesis that ALS-PDC among the Chamorro people of Guam is associated with traditional feasting on flying foxes.

In the editorial accompanying these two articles, Andrew Feigin focuses attention on the need to have better symptomatic treatments for patients with advanced PD. He notes that animal model data have supported the ability of adenosine A2A receptor antagonists to enhance dopamine benefit without worsening dyskinesias. Whereas the use of subthalamic nucleus deep brain stimulation has raised the standard for medical therapies in advanced PD, the results of these two trials are encouraging. Moreover, adenosine A2A antagonists may have the additional benefit of being neuroprotective.
Central Horner’s syndrome and contralateral ataxic hemiparesis

Rossetti et al. studied nine patients with a crossed syndrome of the rostral brainstem: a central Horner’s syndrome associated with contralateral ataxic hemiparesis. This pattern was related to lesions at the paramedian diencephalic-mesencephalic junction. Clinically the disorder has to be differentiated from the more common internal carotid artery dissection, which can produce similar findings.

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Ischemic lesion in the right paramedian thalamic territory, extending to the rostral paramedian mesencephalon (DW-MRI).

Polymyositis (PM): an overdiagnosed entity?

Based on clinical, laboratory, and histopathologic criteria, van der Meulen et al. diagnosed 165 patients with myositis (inclusion body myositis excluded) as follows: polymyositis 9, dermatomyositis 59, unspecified myositis (perimysial/perivascular infiltrates, no PM or DM) 65, possible myositis (necrotizing myopathy, no inflammatory infiltrates) 32. At follow-up, five of the nine patients with PM had typical features of inclusion body myositis.

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Epilepsy risk factors and cortical dysplasia

Patients with medically refractory temporal lobe epilepsy (TLE) frequently have risk factors for epilepsy such as febrile convulsions (FC). Porter et al. found that the majority of children with refractory TLE and a history of FC, who were treated with surgery, actually had cortical dysplasia in temporal neocortex.

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Inflammatory change in myopathy with GNE mutations

Yabe et al. reported two siblings with distal myopathy with rimmed vacuoles (DMRV) with GNE mutations. Inflammation, uncommon in DMRV, was observed in the connective tissue and between muscle fibers.

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The editorial by Amato and Griggs, which accompanies the articles by van der Meulen et al. and Yabe et al., notes that polymyositis remains a diagnosis of exclusion. The list of exclusions continues to lengthen. As newly recognized diseases such as dysferlinopathies and GNE mutations are also found to present with a polymyositis phenotype, polymyositis as a separate disease entity may become rare or cease to exist.

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ID migraine™: A screener for improving migraine recognition

Lipton et al. developed and validated a three-item migraine screener in 563 patients with headache presenting for routine primary care. Those who had two of three cardinal migraine features on the screener (headache-related disability, nausea, and photophobia) had a 93% chance of having a migraine diagnosis assigned by an independent headache expert. The simplicity and brevity of the screener, when coupled with its reliability, sensitivity, and specificity, suggest that it could improve migraine recognition.

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